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MODERN CONCEPTS OF CARDIOVASCULAR DISEASE

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Roentgenologic Considerations in the Diagnosis of Congenital Heart Disease (II)*

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CONDITIONS CHARACTERIZED BY DECREASED PULMONARY VASCULATURE

Decreased pulmonary arterial vasculature is seen in patients who have obstruction to pulmonary arterial flow. This obstruction may be at the pulmonary valve, in the pulmonary outflow tract of the right ventricle, or at the level of the tricuspid valve. In certain rare instances, it may be in the pulmonary artery itself or one or more of its branches. The majority of lesions in this group are associated with a right-to-left shunt through a septal defect, with cyanosis. It is perhaps in this group of anomalies, and in the various types of transposition and truncus discussed in the previous group, that angiocardiography is most valuable.

The most common malformation in this group is pulmonary stenosis with ventricular septal defect, or so-called tetralogy of Fallot. Less commonly seen are cases of pulmonary stenosis with intact ventricular septum, pulmonary atresia, tricuspid atresia, Ebstein's malformation of the tricuspid valve, peripheral pulmonary artery stenosis, and agenesis of a main branch of the pulmonary artery.

Tetralogy of Fallot

Figure 9 shows the findings usually described as characteristic of tetralogy of Fallot, namely, so-called coeur-en-sabot configuration with elevation of the cardiac apex, clear lung fields with flat or concave pulmonary artery segment, and right-sided aortic arch. The bizarre appearance of the vessels present in the lung fields is due to a large collateral flow through the bronchial arteries. These dilated bronchial arteries are easily recognized as short vessels which run in a random, disorderly fashion and

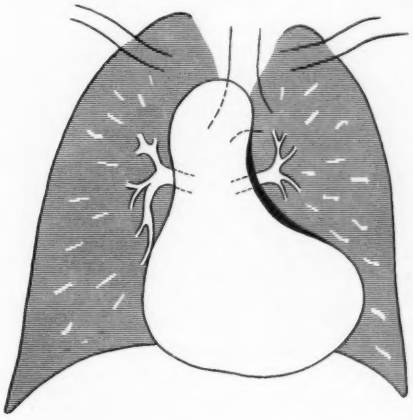

Figure 9. *Tetralogy of Fallot. Note so-called coeur-en-sabot configuration with elevation of apex. Pulmonary artery segment is concave and peripheral vasculature is decreased. Dilated bronchial arteries are present. Aortic arch is prominent and on the right.*

* Part I of this article appeared in the May, 1961, issue.

do not taper as do pulmonary arteries. Unfortunately, from a diagnostic standpoint, the roentgenographic manifestations of tetralogy of Fallot are variable, because the degree of pulmonary stenosis is variable, and this so-called typical picture is seen only in about 30 per cent of cases. In many cases of tetralogy, the pulmonary vasculature appears normal, and in a small percentage, it may even be increased. This increased vasculature is usually not marked, however, and occurs in those patients with so-called pink tetralogy who have only mild pulmonary stenosis, a left-to-right shunt, and no cyanosis. A right-sided aortic arch is more commonly associated with tetralogy than with any other cardiac anomaly except persistent truncus arteriosus, being present in about 25 per cent of cases.

Pulmonary Stenosis with Intact Ventricular Septum

Pulmonary stenosis with intact ventricular septum, often referred to as isolated pulmonary stenosis or pulmonary stenosis with normal aortic root, is a rarer anomaly than tetralogy. In about one half of the cases of pulmonary stenosis with intact ventricular septum, an atrial septal defect or a patent foramen ovale is present. This particular combination of anomalies is sometimes referred to as the trilogy of Fallot. If the pulmonary stenosis is severe enough in such cases, the elevated right ventricular and atrial pressures will produce a right-to-left interatrial shunt, with resultant cyanosis. The pulmonary stenosis is almost always valvular, rather than infundibular as it is in tetralogy, although secondary hypertrophy of the right ventricular outflow tract with significant infundibular stenosis is frequently present as well.

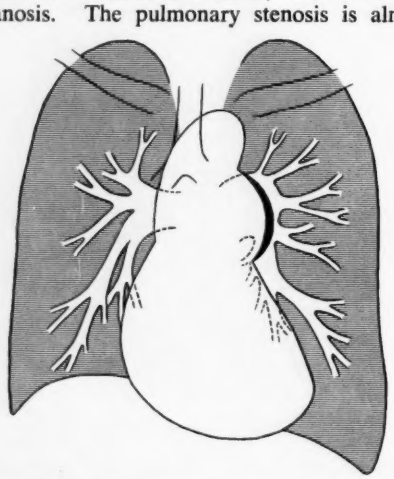

Figure 10. *Pulmonary stenosis with intact ventricular septum. Only abnormal finding is poststenotic dilatation of the pulmonary artery and its left main branch.*

Pulmonary stenosis with intact ventricular septum presents fairly consistent and characteristic findings on the plain roentgenogram, and the radiologist is able to suggest the correct diagnosis in a relatively high percentage of cases (fig. 10). Poststenotic dilatation of the pulmonary artery is a classic roentgenographic finding in this condition, being present in more than 90 per cent of cases. Roentgenographic evidence of poststenotic dilatation is most likely to be absent in patients less than two years of age, and even when present in this age group, it may not be detectable on the roentgenogram if the thymus is prominent. The poststenotic dilatation usually is localized to the main pulmonary artery, but in some instances the left main branch of the pulmonary artery also is dilated, the right main branch appearing normal. Fluoroscopically, the dilated pulmonary artery always shows increased pulsation, and in most instances this increased pulsation is fairly marked.

Although in most instances of pulmonary stenosis with intact ventricular septum the peripheral pulmonary vasculature is normal, in about 20 per cent of patients the vasculature is decreased from normal. Decreased vasculature is usually seen in infants who have severe pulmonary stenosis with an associated interatrial communication and a right-to-left shunt. Poststenotic dilatation of the pulmonary trunk in these patients with decreased vasculature, when present, serves to differentiate them from patients with tetralogy, pulmonary atresia, tricuspid atresia, and Ebstein's malformation, all of which show decreased pulmonary vasculature but are characterized by a flat or concave pulmonary artery segment. Right ventricular enlargement is a fairly consistent finding in functionally significant pulmonary stenosis, as is enlargement of the right atrium.

Pulmonary Atresia

Pulmonary atresia may occur with ventricular septal defect (pseudotruncus) or, rarely, with intact ventricular septum. In pulmonary atresia with ventricular septal defect, the blood supply to the lungs occurs in one of two ways: either by means of large bronchial arteries arising from the descending aorta, or by means

of a patent ductus which communicates with intact pulmonary arteries. Pulmonary atresia with ventricular septal defect may be looked upon as being only a severe form of tetralogy.

There are two types of pulmonary atresia with intact ventricular septum. An atrial septal defect is always present in these cases. In one type, the right ventricular cavity is large, and blood escapes from the right ventricle only through an incompetent tricuspid valve. In the other type, the right ventricular cavity is quite small, and the tricuspid valve is competent. In some of these cases, blood escapes from the right ventricle by means of persistent right ventricular myocardial sinusoids which communicate with the coronary vessels. Blood reaches the lungs, then, from the aorta, either by means of bronchial arteries or a patent ductus. Patients with pulmonary atresia and intact ventricular septum have severe symptoms at birth and usually do not survive more than a few weeks.

The pulmonary vasculature in pulmonary atresia is greatly decreased. In pulmonary atresia with ventricular septal defect (pseudotruncus), the aortic arch is on the right in approximately 25 per cent of cases. Roentgenographically, this condition often cannot be distinguished from some of the other anomalies producing obstruction to pulmonary outflow, such as tetralogy and tricuspid atresia (fig. 11). The differential diagnosis of these malformations often can be made with accuracy only by means of angiocardiography.

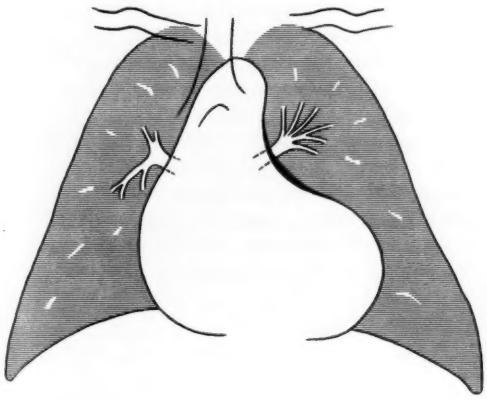

Figure 11. *Tricuspid atresia. Except for presence of small left aortic arch, similarity to case of tetralogy represented in figure 9 is striking.*

Tricuspid Atresia

In this condition, there must always be an associated atrial septal defect; blood from the right atrium flows through the defect into the left atrium, then into the left ventricle. From the left ventricle, it may reach the lungs by way of a ventricular septal defect and a rudimentary right ventricle and pulmonary artery, or by bronchial arteries arising from the aorta, or by way of a patent ductus. Decreased pulmonary vasculature and a concave pulmonary artery segment are rather consistent findings in tricuspid atresia (fig. 11). As discussed above, the roentgenographic findings in tricuspid atresia are often indistinguishable from those of severe tetralogy of Fallot and of pulmonary atresia. Severe pulmonary stenosis with intact septum in infants who do not have poststenotic dilatation of the pulmonary artery may also produce an identical picture. A right-sided aortic arch, however, in tricuspid atresia or pulmonary stenosis with intact septum is practically unknown.

Ebstein's Malformation

Ebstein's malformation of the tricuspid valve is another condition to be considered in this group. This anomaly consists of deformity and distal displacement of part of the tricuspid valve into the right ventricle. Although the tricuspid-valve ring may be normally situated, the valve leaflets partially arise from the right ventricular wall and ventricular septum and are fused into a membranous structure which extends like a basket or curtain down into the cavity of the right ventricle. Part of the right ventricular cavity, therefore, is anatomically and functionally a part of the right atrium, and the functioning right atrium is grossly enlarged. That part of the right ventricle which functions as such is small. Because the tricuspid orifice is usually stenotic, as well as incompetent, and the diastolic capacity and stroke volume of the functioning right ventricle are reduced, pulmonary blood flow is diminished. A patent foramen ovale or atrial septal defect is commonly present, and there may be a right-to-left shunt through this defect, with cyanosis.

Although there is nothing pathognomonic about the roentgenographic findings in Ebstein's malformation, a greatly enlarged, globular heart, with a flat pulmonary artery segment and clear lung fields, is consistently seen (fig. 12). With experience, the radiologist

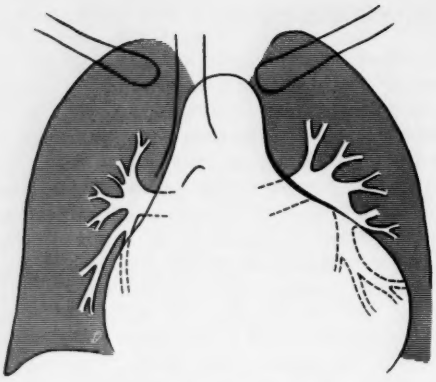

Figure 12. *Ebstein's malformation. Note large, globular heart. Pulmonary artery segment is flat, or slightly concave, and peripheral vasculature is slightly decreased.*

can suggest the diagnosis of Ebstein's malformation with considerable accuracy from the appearance of the posteroanterior roentgenogram.

Congenital Absence of Main Branch of Pulmonary Artery

Congenital absence of either the right or the left main branch of the pulmonary artery is a relatively rare condition which produces unilateral decrease in the pulmonary arterial vasculature. Characteristically, the hilar and peripheral branches of the pulmonary artery cannot be visualized on the involved side, but dilated bronchial arteries are usually visible. There is frequently compensatory hypertrophy of the opposite main branch of the pulmonary artery, and the thorax on the involved side is small, with shift of the mediastinal structures to this side (fig. 13). Whenever these findings are encountered, congenital absence of a main branch of the pulmonary artery is likely. This condition is always associated with hypoplasia or agenesis of some of the segments of the involved lung. Angiocardiography and bronchography are necessary for an exact anatomic diagnosis.

CONDITIONS WITH NORMAL PULMONARY VASCULATURE

The common, operable lesions in this group are coarctation of the aorta and congenital aortic stenosis. It is important to recognize, of course, that many of the lesions included in other groups, and already discussed, are many times associated with normal pulmonary vasculature.

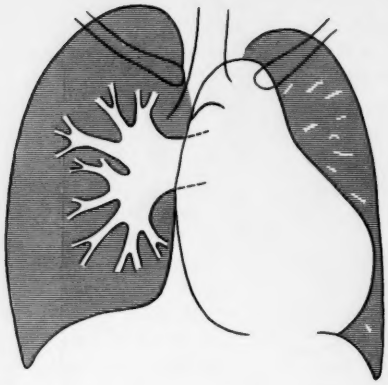

Figure 13. *Agenesis of left main branch of the pulmonary artery. Heart and mediastinal structures are shifted to the left, and the left hemithorax is small. No pulmonary vessels are visible in the left lung, but dilated bronchial arteries are present. There is compensatory enlargement of right main branch of the pulmonary artery.*

Coarctation of Aorta

Coarctation of the aorta may be diagnosed from the conventional roentgenogram of the thorax in about 85 per cent of patients with this anomaly. The characteristic features of coarctation are present in the diagram shown in figure 14; there is bilateral rib notching and the actual site of the coarcted segment can be seen, producing the characteristic figure-of-3 conformation of the descending arch of the aorta. The dilated left subclavian artery is commonly seen when the coarctation is distal to the origin of this vessel. Also, the left ventricle is enlarged. Rib notching is present in about 80 per cent of patients with coarctation, and the site of the coarcted segment can be visualized in approximately 25 per cent of cases. In infants and children, these characteristic signs are not present, and the diagnosis of coarctation must rest on clinical evidence and demonstration of the lesion by angiocardiography or thoracic aortography.

In rare instances, unilateral rib notching may occur in coarctation and, when present, gives important information regarding the site of the coarctation and its relation to the branches of the aortic arch. The presence of rib notching on the right side alone indicates that the site of the coarctation is proximal to the origin of the left subclavian artery. When rib notching occurs in coarctation on the left side only, this indicates the presence of an anomalous right subclavian artery arising from the de-

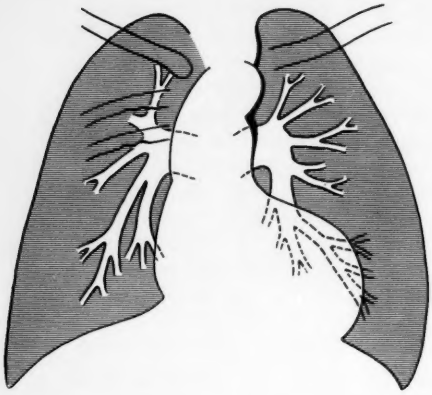

Figure 14. *Coarctation of the aorta. Left ventricle is enlarged. Ascending aorta and arch are prominent. Note figure-of-3 conformation of descending aorta at site of coarctation. Left subclavian artery is dilated, and rib notching is present.*

scending aortic arch distal to the site of the coarctation.

Congenital Aortic Stenosis

The roentgenologic manifestations of congenital aortic valvular stenosis are similar to those of acquired aortic valvular stenosis. Findings consist of a varying degree of left ventricular enlargement and calcification of the aortic valve. While calcification of the valve is rarely, if ever, seen in children, such calcification is not uncommonly encountered in adults with congenital aortic valvular stenosis. Another common finding is poststenotic dilatation of the ascending aorta, which may be seen best on the posteroanterior and the left anterior oblique roentgenogram. In subvalvular and supravalvular aortic stenosis, enlargement of the left ventricle is often the only abnormal roentgenologic sign present.

CONDITIONS CAUSING PULMONARY VENOUS HYPERTENSION (OBSTRUCTION TO PULMONARY VENOUS FLOW)

Obstruction to venous return from the lungs may occur at the left ventricular level, the mitral valve level, or the pulmonary vein level. Obstruction at the left ventricular level occurs in left ventricular failure from any cause. The most important operable congenital anomalies, however, that produce obstruction to pulmonary venous flow are congenital mitral stenosis and congenital stenosis of the pulmonary veins.

Congenital Mitral Stenosis

When congenital mitral stenosis occurs as an isolated lesion, the roentgenologic findings are the same as those in acquired mitral stenosis. In older patients with mitral stenosis with marked left atrial enlargement or calcification of the mitral valve, and with the classic pulmonary signs of chronic pulmonary venous hypertension, diagnosis is not difficult. However, in early mitral stenosis in which the only abnormal findings are slight left atrial enlargement and engorgement of the hilar vessels, differentiation from a congenital anomaly with pulmonary hypertension, such as septal defect, may seem to offer some difficulty. In mitral stenosis, however, the appearance of the pulmonary vessels is different from that seen in patients with congenital heart disease and pulmonary hypertension due to increased pulmonary artery flow. For example, in a patient with atrial septal defect and increased pulmonary artery flow, the individual outlines of the engorged pulmonary arteries can be well seen extending into the periphery of the lung fields. In this condition, there is only pulmonary arterial hypertension and no pulmonary venous hypertension. In cases of mitral stenosis, the prominent hilar vessels and the lung fields have a hazy, amorphous appearance and the outlines of individual vessels do not show as well. This is due to the fact that in mitral stenosis there is not only pulmonary arterial hypertension but also pulmonary venous hypertension, with engorgement of veins, lymphatics, and arteries. This hazy appearance of the pulmonary vasculature is best noted in the right paracardiac region, since this is where the engorged right pulmonary veins cross medially just below the pulmonary arteries before entering the left atrium. In a patient with atrial septal defect and pulmonary hypertension, this area is relatively clear, since the veins and lymphatics are not distended.

Another important differentiating feature is the pulsation of the hilar arteries as seen fluoroscopically. Pulmonary arterial hypertension in mitral stenosis occurs as a passive or secondary phenomenon. In other words, when the pulmonary venous pressure increases, the pulmonary arterial pressure rises secondarily in order to maintain a certain pressure gradient across the pulmonary capillary bed, and not because of increased pulmonary flow. A poorly understood vasoconstrictive reflex mechanism, causing constriction of the peripheral pulmo-

nary arteries, also plays a major role in the production of pulmonary arterial hypertension in mitral stenosis. Thus, in mitral stenosis, the pulmonary arteries usually do not show increased pulsations, and their pulsations are often diminished from normal. In septal defect with increased pulmonary artery flow, the pulmonary artery pulsations are, of course, increased.

Congenital Stenosis of Pulmonary Veins

Congenital stenosis of the pulmonary veins is a rare anomaly that produces severe and rather acute pulmonary venous hypertension, usually evident soon after birth. Although this anomaly is rare, it is important in that its early recognition could be lifesaving. The roentgenographic findings, frequently characteristic, consist of a cardiac silhouette of fairly normal size and configuration, and congestive changes in the lungs ranging from simple distention of pulmonary veins to frank pulmonary edema. At the Mayo Clinic, we have encountered only a few patients with stenosis of the pulmonary veins, but the features shown in the diagram in figure 15, although perhaps a little more striking than usual, have been characteristic of most of these cases. These pulmonary changes may be unilateral when only the pulmonary veins on one side are involved or may involve only one lobe of a lung when the pulmonary venous stenosis is limited to one pulmonary vein.

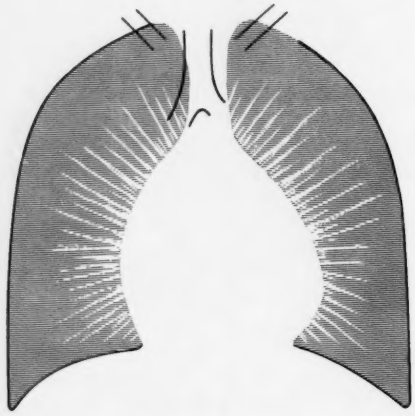

Figure 15. *Congenital stenosis of the pulmonary veins. Heart is normal in size and contour. Appearance of pulmonary edema is due to presence of acute, severe pulmonary venous hypertension.*

Obstruction to pulmonary venous flow may also be seen in patients with partial or total anomalous venous connection to the inferior vena cava or hepatic veins below the diaphragm or, more commonly, when there is anomalous venous connection into the portal vein; in the latter instance, the pulmonary venous hypertension is due to the fact that the venous blood from the lungs must pass through the liver before returning to the right side of the heart, as well as to the fact that the anomalous vein or veins may be compressed by the action of the diaphragm.

SUMMARY

Roentgenologic examination plays an important role in the diagnosis and surgical evaluation of congenital malformations of the heart and great vessels, and an orderly approach to analysis of the roentgenologic findings in this group of diseases is to be stressed. Such an approach, if the method is to achieve its maximal usefulness, must be based on a fundamental understanding of the pathology and altered hemodynamics of congenital heart disease.

A classification of congenital anomalies into four groups, based on changes in the lesser circulation, and further subdivision, based on certain other differentiating features, has proved to be of great practical value from the standpoint of roentgenologic interpretation.

Although, in most instances, the roentgenologic findings alone are not indicative of a specific cardiac anomaly, by the use of this suggested approach and correlation of the roentgenologic findings with the clinical findings in a given patient, conventional roentgenography may prove to be an important diagnostic aid. In many instances, of course, specialized procedures, such as cardiac catheterization, angiocardiography, and thoracic aortography, must be employed if a precise diagnosis is to be reached.